

Frequently Asked Questions

Variant Creutzfeldt-Jakob Disease (vCJD)

What is Variant Creutzfeldt-Jakob Disease (vCJD)?

- Variant Creutzfeldt-Jakob Disease (vCJD) is a rare, fatal, degenerative brain disease caused by abnormal, transmissible proteins called prions.

How common is vCJD?

- vCJD is very rare; there has never been a case of vCJD reported in Utah.
- Countries that have reported cases of vCJD include Great Britain, France, Canada, Ireland, Hong Kong, and Italy. The United States has reported two cases, however both persons were believed to have acquired the infection outside of the United States; there has never been a reported case of vCJD that was contracted in the United States.

How is vCJD transmitted?

- It is thought that vCJD is acquired from eating meat products containing parts of the nervous system from cows with a related disease.
- vCJD may also be transmitted through blood transfusions using blood products from an infected donor.
- Transmission of vCJD from one person to another person through direct contact has not been documented.

Who can get vCJD?

- Most cases are identified in individuals between 16 and 45 years of age; the average age of diagnosed cases is 29 years.

What are the symptoms of vCJD?

- The first symptoms of vCJD often include anxiety, depression (including absence of emotion or lack of interest in surroundings, weight loss, and insomnia), withdrawal, and behavior changes, so patients are often referred first to a psychiatrist rather than a neurologist when symptoms develop.
- Often painful sensory symptoms are the first neurological symptoms to develop.
- Several weeks or months after initial onset of symptoms, development of sudden jerky movements may occur.
- Anxiety, depression, withdrawal and behavioral changes may persist.
- Progressive dementia (including loss of mental function marked by symptoms such as memory loss) may develop.
- Persistent pain and odd sensations in the face and limbs may occur.
- Eventually the patient may lose the ability to move or speak and will need constant nursing care.
- Death usually occurs approximately one year after onset of symptoms.

What is the incubation period for vCJD?

- Based on available information and suspected exposures for vCJD, incubation period for the disease is estimated to be 1 to 30 years.

How is vCJD diagnosed?

Frequently Asked Questions

Variant Creutzfeldt-Jakob Disease (vCJD)

- After symptoms appear, doctors can often make a preliminary diagnosis based on medical history, physical examination (including of the neurological system), and certain diagnostic tests including:
 - Electroencephalogram (EEG)
 - Magnetic resonance imaging (MRI)
 - Spinal fluid tests
 - Tonsil biopsy
- A confirmed diagnosis of vCJD can usually only be made after the patient's death.

How is vCJD treated?

- Symptoms of the disease are treatable, but there is no treatment available that slows or stops the disease.

Is there a vaccine or medicine available that cures or prevents vCJD?

- No vaccine is currently available to prevent vCJD, and no medicines have been identified to cure or prevent the disease.

What is being done to prevent vCJD?

- Research is being done to better determine what causes vCJD and identify possible treatments for the disease.
- To ensure that the blood supply is safe in the United States, in 1999 the FDA began to exclude blood donations from people who have lived in countries identified as having a risk for prion diseases.
- Also, in the United States the FDA and USDA work together to protect the food supply. Protection steps that have been implemented to prevent transmission of vCJD include:
 1. In 1989 a ban was put in place on importing live ruminants (cows, sheep, and goats), and higher controls were put in place on ruminant products from countries with possibly diseased animals.
 2. In 1990 the USDA began a surveillance program testing approximately 20,000 cows per year for disease. If a diseased cow is found there is an emergency response plan in place to trace the source of disease for the cow and implement appropriate control measures to limit human exposure to possibly contaminated products.
 3. In 1997 the FDA put a Feed Rule in place that includes:
 - Banning most protein from mammals in ruminant animal feed.
 - A mandatory inspection procedure for companies that handle animal feed to ensure compliance with the Feed Rule.
 4. In 2003 the USDA added more safeguards including:
 - Banning all material from cattle that cannot walk (downer cattle) from the human food supply.
 - Holding all products from cattle tested for disease until the cattle are confirmed to be disease-free.
 - Prohibiting the use of cow brains, spinal cords, certain organs, and other possibly prion-infected material from cattle older than 30 months in human food.
 - Prohibiting the use of the small intestine from cattle in human food.
 - Prohibiting the scraping of muscle tissue away from bones of dead cows. This ban will

Frequently Asked Questions

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ensure that spinal cord and other high-risk tissues are not included in products for human use.

- To date, only two cows have been found in the United States that were positive for the prion illness related to human vCJD. Both cows were from Canada and did not enter the human food chain.

Is vCJD reportable in Utah?

- Both classic CJD and vCJD should be reported to your local health department or the Utah Department of Health.

How is vCJD different than Classical CJD?

	Classical CJD	Variant CJD
Found in the United States	Yes	No
Possibly food-related	No (insufficient evidence)	Yes (suggested bovine-to-human)
Average age of patient at time of diagnosis	60 years	29 years
Disease duration until death	Average is 5 months; 90% of reported cases die in less than 1 year	Average is 14 months
Possible genetic link	Yes	No

Where can people get more information?

- More information is available from your physician, the Creutzfeldt-Jakob Disease Foundation (330-665-5590) www.cjdfoundation.org, the CJD Surveillance National Prion Disease Pathology Surveillance Center (216-368-0587) www.cjdsurveillance.com, your local health department, the Utah Department of Health (801-538-6191), or the Centers for Disease Control and Prevention (CDC) <http://www.cdc.gov/ncidod/dvrd/cjd/>.